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As previously stated (2), we believe that coronary angiography should be part of the diagnostic work-up in those children with HCM who have clinical indicators of ischemia. We have used surgical unroofing of the myocardial bridge without complication (2). All patients undergoing this procedure have demonstrated a marked change in their clinical outcome with evidence of reversal of ischemia (2).

Coronary stenting has not been previously used in children with HCM. Given the authors' statements concerning high rates of restenosis, we find it curious that they consider this an attractive therapeutic strategy.

In summary, children with HCM carry an increased risk of sudden death. There is evidence to suggest that myocardial ischemia may be the cause of death in these patients and may occur from several causes including myocardial bridging (2). Myocardial bridging is a treatable cause of myocardial ischemia (2,5–7), which may prove to be a life-saving therapy in these patients.

We agree with the authors in that the association between myocardial bridging and sudden death in patients with HCM is a potentially important finding, and as such we believe that this warrants further study with a multicentered prospective trial.

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REFERENCES

1. Yetman AT, Hamilton RM, Benson LN, McCrindle BW. Long-term outcome and prognostic determinants in children with hypertrophic cardiomyopathy. J Am Coll Cardiol 1998;32:1943–50.
2. Yetman AT, MacDonald C, McCrindle BW, Freedom RM, Gow RM. Myocardial bridging with coronary compression in children with hypertrophic cardiomyopathy: a risk factor for sudden death. N Engl J Med 1998;339:2101–9.
3. Sadoul N, Prasad K, Elliott PM, Bannerjee S, Frenneaux MP, McKenna WJ. Prospective prognostic assessment of blood pressure response during exercise in patients with hypertrophic cardiomyopathy. Circulation 1997;96:2987–91.
4. Yoshida N, Ikeda H, Wada T, et al. Exercise-induced abnormal blood pressure responses are related to subendocardial ischemia in hypertrophic cardiomyopathy. J Am Coll Cardiol 1998;32:1938–42.
5. Tio Ra, Van Gelder IC, Boonstra PW, Crijns HJGM. Myocardial bridging in a survivor of sudden cardiac near-death: role of intracoronary Doppler flow measurements and angiography during dobutamine stress in the clinical evaluation. Heart 1997;77:280–2.
6. Agirbasli M, Martin GS, Stout JB, Jennings HS, Lea JW, Dixon JH. Myocardial bridge as a cause of thrombus formation and myocardial infarction in a young athlete. Clin Cardiol 1997;20:1032–6.
7. Pey J, de Dios RM, Epeldegui A. Myocardial bridging and hypertrophic cardiomyopathy: relief of ischemia by surgery. Int J Cardiol 1985;8:327–30.

Electrical Storm in Patients With Transvenous Implantable Cardioverter-Defibrillators

We read with interest the paper by Credner et al. (1) reporting an incidence of what is called "electrical storm" or clustering of arrhythmia episodes in patients with implantable cardioverter-defibrillators (ICDs). This report is similar to other reports suggesting a 10% incidence of this phenomenon among ICD recipients. Credner et al. reported precipitating factors for arrhythmia episodes in only 26% of patients, and the patients with electrical storm had a trend toward having a reduced baroreflex slope.

Our concern is that reports of arrhythmia clustering, such as the one from Credner et al. and others, fail to take into account fundamental observations on the underlying temporal patterns of arrhythmia recurrence (1,2). The concept of an electrical storm is intended to designate a marked increase in arrhythmia frequency that deviates from a baseline frequency. An understanding of this baseline frequency or pattern is necessary to define electrical storm, yet is rarely considered. This leaves the definition of electrical storm completely arbitrary. Previous work has demonstrated that recurrent ICD detections are nonrandomly distributed over long-term follow-up in most patients (90%) we have studied (3). Further analysis of these episodes has shown that there is a distinct tendency for clustering of arrhythmia recurrences during short periods. In fact, when considering the interval between consecutive ICD detections in 31 patients, 55% of these intervals had a value of ≤1 h. Thus, two episodes of ventricular arrhythmias within 1 h is not unusual. Although the concept of electrical storm certainly has validity for extreme cases, the definition of a storm as ≥2 to 3 episodes in 24 h, as in previous studies, is overly inclusive (2,3). More recently, we have demonstrated a fractal pattern to the distribution of episodes of ventricular tachycardia in patients with frequent ICD detections (4). This pattern again indicates that short intervals between tachycardia episodes are much more likely than long intervals between episodes, and that both the long and short intervals are part of a single consistent power law distribution.

These data lead us to believe that the recurrences of ventricular tachyarrhythmias in patients with an ICD are not random, but follow a discernable pattern. This tendency toward clustering of arrhythmia recurrences must be considered when defining electrical storm. Because these events form one distribution, comprising both the long and short intervals, the criteria for electrical storm based on a number of events within a brief interval are arbitrary and do not reflect the true nature of the underlying distribution. A better index of deviation from the expected pattern would appear to be the relative number of short to long intervals that are related to the fractal dimension. Arrhythmia recurrence is likely to depend on the convergence of several physiologic conditions that may produce long-lasting (hours to days) changes in the cardiac milieu that are conducive to the occurrence of ventricular arrhythmias. Further definition of the temporal pattern of arrhythmia recurrences may lead to a better understanding of the complex mechanisms of arrhythmia induction. In addition, defining the efficacy of suppressive antiarrhythmic therapy will benefit from an understanding of the pattern of arrhythmia recurrences. The definition of electrical storm must therefore be derived from an understanding of the "expected pattern" of arrhythmia recurrence. The long-term temporal pattern of arrhythmia recurrence is a neglected area of research and warrants further investigation.

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REFERENCES

1. Credner SC, Klingenheben T, Mauss O, Sticherling C, Hohnloser SH. Electrical storm in patients with transvenous implantable cardioverter-defibrillators. J Am Coll Cardiol 1998;32:1909–15.
2. Dorian P, Cass D. An overview of the management of electrical storm. Can J Cardiol 1997;13:13A–7A.
3. Wood MA, Simpson PM, Stambler BS, Herre JM, Bernstein RC, Ellenbogen KA. Long-term temporal patterns of ventricular arrhythmias. Circulation 1995;91:2371–7.
4. Liebovitch LS, Todorov AT, Zochowski M, et al. Nonlinear scaling properties of cardiac rhythm abnormalities. Phys Rev E 1999;59:3312–9.

REPLY

We appreciate the interest of Wood et al. in our recently published paper on electrical storm in patients with implantable cardioverter-defibrillators (ICDs) (1). In their comments, they reemphasize the importance of defining electrical storm based on temporal patterns of arrhythmia clustering. As stated in our report (1), the precise definition of this syndrome is still evolving; therefore, we adhered to the most commonly used definition (2,3), realizing that it is still somewhat arbitrary. Specifically, we are aware of the findings of Wood et al. (4) indicating that two episodes of arrhythmia detection by the ICD within 1 h may not necessarily be an unusual finding. It appears important to note, however, that the median number of arrhythmic episodes constituting electrical storm in our patients was 17 within a single 24-h period leading to the need for urgent therapy in most patients (1). Although we had to focus on the timing of arrhythmia clustering, as with any definition of electrical storm, the clinical picture substantially contributed to the definition in our series. Moreover, we were interested in several other issues related to electrical storm, such as precipitating factors, therapeutic measures and potential prognostic implications. In summary, we agree with Wood et al. (4) that more research is needed to arrive at a more precise definition of electrical storm. Our paper aimed to be one step in this direction.

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REFERENCES

1. Credner SC, Klingenheben T, Mauss O, Sticherling C, Hohnloser SH. Electrical storm in patients with transvenous implantable cardioverter-defibrillators. J Am Coll Cardiol 1998;32:1909–15.
2. Kowey PR, Levine JH, Herre JM, et al. Randomized, double-blind comparison of intravenous amiodarone and bretylium in the treatment of patients with recurrent, hemodynamically destabilizing ventricular tachycardia or fibrillation. Circulation 1995;92:3255–63.
3. Scheinman MM, Levine JH, Cannom DS, et al. Dose-ranging study of intravenous amiodarone in patients with life-threatening ventricular tachyarrhythmias. Circulation 1995;92:3264–72.
4. Wood MA, Simpson PM, Stambler BS, Herre JM, Bernstein RC, Ellenbogen KA. Long-term temporal patterns of ventricular arrhythmias. Circulation 1995;91:2371–7.

Use of Radial Artery Applanation Tonometry

Cameron et al. (1) reported clinical evaluation of a system that was developed at St. Vincent's Hospital by us (2) and that uses a generalized transfer function to estimate the calibrated ascending aortic pressure waveform from the radial artery pressure pulse wave. The authors concluded that waveform analysis is of limited value and that simple linear relations are sufficient to generate central from cuff sphygmomanometric pressure values in individual patients. We disagree.

In our own continuing evaluation, we have interrogated our large data base (15,533 reports in 1,604 patients/subjects) and have participated in studies where the estimated calibrated ascending aortic waveform is compared to the pressure wave recorded simultaneously by an intraarterial catheter from the ascending aorta (3–5). In the first group of studies, we have obtained results for systolic, diastolic and augmented pressure, which are very similar to those reported by Cameron et al., and have shown the same wide scatter in values of systolic and augmented pressure for measured radial and estimated aortic waveforms. We take these results to show the potential for generating more precise indexes of left ventricular load and function than those available with the cuff sphygmomanometer alone. In the second group of studies, we have compared directly measured and estimated ascending aortic pressure waves and indexes derived therefrom. We have shown a close correspondence between estimated and measured aortic pressure indexes in individual patients under control conditions and with physiologic (Valsalva maneuver) and pharmacologic perturbations. Indeed, correspondence between estimated and measured ascending aortic measurements generally fell within the AAMI (Association for the Advancement of Medical Instrumentation) requirements for comparing different methods (6), whereas correspondence between measured aortic and upper limb values did not. Virtually identical results have been reported for a similar system (7,8).

The evaluation by Cameron et al. (1) downplays the value of information carried by the arterial pressure waveform and focuses on cuff sphygmomanometric values. Their evaluation is limited by the fact that all measurements (brachial cuff sphygmomanometry with radial tonometry) were taken in the upper limb and none from a central artery. Their comparison of "central" and peripheral mean pressure was simply of the integrated calibrated radial waveform against the mean brachial value determined by an oscillometric method.

We remain convinced that use of the cuff sphygmomanometer can be improved by incorporation of information provided by the pulse waveform in the upper limb. Studies such as that by